

## **Prune Belly Syndrome**

Aka Eagle-Barrett syndrome or triad syndrome

*Failed abdominal wall development*

*Bilateral intra-abdominal testes*

*Urinary tract anomalies*

### Demographics

1:30,000 live births

Males >> females (95%)

Twins > singletons

Unknown aetiology – multiple theories

Possibly temporary glanular urethral obstruction, leading to dilated urethra and bladder. Massively enlarged bladder proposed to inhibit normal development of abdo wall musculature

Associated with trisomy 18 and 21

### Clinical features

Failed abdominal wall development

Ranges from total absence to limited defects

Medial and inferior elements most commonly absent

Bilateral intra-abdominal testes

Most commonly at level of CIA

Histology similar to non-PBS patients

Spontaneous paternity never reported

ICSI successful in a few case reports

Urinary tract anomalies

Renal dysplasia

50%, determines severity

Hydroureteronephrosis

lack of ureteric smooth muscle, distal > proximal

Massive bladder

pseudodiverticulum at dome

patent urachus in 25%

increased ratio of collagen to muscle

50% void spontaneously with normal UDS and low PMRs

Posterior urethra

Dilated prostatic urethra

Hypoplastic prostate

Dilated, occasionally absent vas and seminal vesicles

Typically retrograde ejaculation

Anterior urethra

Megalourethra

Urethral atresia (fatal unless patent urachus)

Associated non-urological abnormalities

Overall seen in ~75%

Pulmonary – hypoplasia, pneumothorax (50%)

Orthopaedic - scoliosis, congenital hip dislocation (50%)

Gastrointestinal - malrotation, atresia, stenosis, volv.(30%)

Cardiac - PDA, VSD, ASD, tetralogy (10%)

Diagnosis

Classic prenatal USS findings

Hydroureteronephrosis

Distended bladder

Irregular abdominal wall

Typically not seen however. Difficult therefore to distinguish from causes of bladder outflow obstruction such as posterior valves

No correlation between the degree of hydroureteronephrosis and renal dysfunction

Classification (Described by Woodard 1985)

<b>Category</b>	<b>Characteristics</b>
I	Renal dysplasia
	Oligohydramnios
	Pulmonary hypoplasia
	Potter's features
	Urethral atresia
II	Full triad features
	Minimal or unilateral renal dysplasia
	No pulmonary hypoplasia
	May progress to renal failure
III	Incomplete or mild triad features
	Mild to moderate uropathy
	No renal dysplasia
	Stable renal function
	No pulmonary hypoplasia

Category 1 patients almost always either stillborn or die in perinatal period; exception is those patients with patent urachus.

Category 3 patients usually do well. Urologic intervention reserved for those with complications (i.e. UTI)

Category 2 patients intermediate risk. Intervention to prevent renal decline controversial

Management

Initial management in all patients

MDT consisting of neonatology, nephrology, urology plus others

U+E

Renal tract USS

VCUG with ABx cover if renal insufficiency or evidence of BOO.

CXR for pneumothorax, pneumomediastinum, and pulmonary hypoplasia

SPC for BOO

? Circumcision (recommended in Campbell's)

### Orchidopexy

Complete transperitoneal mobilisation with division of vascular pedicles readily achievable if performed at ~6 months  
Alternatively Fowler-Stephens (one or two-stage) or autotransplantation and microvascular anastomosis

### Management of Category 2 patients (PBS and renal insufficiency)

Aim to prevent upper tract UTI

Reduction cystoplasty, ureteric shortening, tapering and reimplantation with or without abdominoplasty (eg. Monfort technique)

Recommended by some with good long-term preservation of renal function (Woodard)

Others advocate careful surveillance with intervention for repeated febrile UTI or renal deterioration (Woodhouse)

### Prognosis

Nadir creatinine in infancy useful predictor of long-term function

If creatinine <60  $\mu\text{mol/l}$  long-term renal function usually stable

30% of patients with impaired renal function develop chronic renal failure before adulthood